



bradyopsia

Bradyopsia is a rare condition that affects vision. The term "bradyopsia" is from the Greek words for slow vision. In affected individuals, the eyes adapt more slowly than usual to changing light conditions. For example, people with this condition are blinded for several seconds when going from a dark environment into a bright one, such as when walking out of a darkened movie theater into daylight. Their eyes also have trouble adapting from bright light to dark conditions, such as when driving into a dark tunnel on a sunny day.

Some people with bradyopsia also have difficulty seeing some moving objects, particularly small objects moving against a bright background. As a result, they often have trouble watching or participating in sports with a ball, such as soccer or tennis. People with bradyopsia can have reduced sharpness (acuity) of vision, although acuity may depend on the conditions under which vision is tested. Visual acuity may appear to be severely affected if it is tested under bright lights, but it can be near normal if tested in a dim environment. The ability to see colors and distinguish between them is normal.

The vision problems associated with bradyopsia become apparent in early childhood. They are usually stable, which means they do not worsen over time.

Frequency

Bradyopsia appears to be rare. Only a few affected individuals worldwide have been described in the medical literature.

Genetic Changes

Bradyopsia can be caused by mutations in the *RGS9* gene or in the *RGS9BP* gene (which is also known as *R9AP*). These genes provide instructions for making proteins that are necessary for normal vision. The proteins are found in light-detecting cells in the eye called photoreceptors. When light enters the eye, it stimulates specialized pigments in these cells. This stimulation triggers a series of chemical reactions that produce an electrical signal, which is interpreted by the brain as vision. Once photoreceptors have been stimulated by light, they must return to their resting state before they can be stimulated again. The *RGS9* and *RGS9BP* proteins play an essential role in returning photoreceptors to their resting state quickly after light exposure.

Mutations in either the *RGS9* or *RGS9BP* gene prevent photoreceptors from recovering quickly after responding to light. Normally they return to their resting state in a fraction of a second, but in people with mutations in one of these genes, it can take ten seconds or longer. During that time, the photoreceptors cannot respond to light. This delay

causes temporary blindness in response to changing light conditions and interferes with seeing small objects when they are in motion.

In some people with bradyopsia, no mutations in the *RGS9* or *RGS9BP* gene have been found. The cause of the condition in these individuals is unknown.

Inheritance Pattern

This condition is inherited in an autosomal recessive pattern, which means both copies of the gene in each cell have mutations. The parents of an individual with an autosomal recessive condition each carry one copy of the mutated gene, but they typically do not show signs and symptoms of the condition.

Other Names for This Condition

- PERRS
- prolonged electroretinal response suppression

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Prolonged electroretinal response suppression
<https://www.ncbi.nlm.nih.gov/gtr/conditions/C1842073/>

Other Diagnosis and Management Resources

- Children's Hospital of Pittsburgh: Electroretinogram
<http://www.chp.edu/our-services/ophthalmology/visual-electrophysiology-diagnostic-lab/electroretinogram>
- MedlinePlus Encyclopedia: Electroretinography
<https://medlineplus.gov/ency/article/003388.htm>
- Prevent Blindness: Living Well with Low Vision
<http://lowvision.preventblindness.org/>

General Information from MedlinePlus

- Diagnostic Tests
<https://medlineplus.gov/diagnostictests.html>
- Drug Therapy
<https://medlineplus.gov/drugtherapy.html>
- Genetic Counseling
<https://medlineplus.gov/geneticcounseling.html>

- Palliative Care
<https://medlineplus.gov/palliativecare.html>
- Surgery and Rehabilitation
<https://medlineplus.gov/surgeryandrehabilitation.html>

Additional Information & Resources

MedlinePlus

- Encyclopedia: Electroretinography
<https://medlineplus.gov/ency/article/003388.htm>
- Health Topic: Vision Impairment and Blindness
<https://medlineplus.gov/visionimpairmentandblindness.html>

Genetic and Rare Diseases Information Center

- Bradyopsia
<https://rarediseases.info.nih.gov/diseases/12299/bradyopsia>

Additional NIH Resources

- National Eye Institute: Diagram of the Eye
<https://nei.nih.gov/health/eyediagram>
- National Eye Institute: Low Vision
<https://nei.nih.gov/health/lowvision/>

Educational Resources

- Disease InfoSearch: Prolonged electroretinal response suppression
<http://www.diseaseinfosearch.org/Prolonged+electroretinal+response+suppression/9172>
- MalaCards: bradyopsia
<http://www.malacards.org/card/bradyopsia>
- Orphanet: Bradyopsia
http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=75374

Patient Support and Advocacy Resources

- American Foundation for the Blind: What is Low Vision?
<http://www.visionaware.org/info/your-eye-condition/eye-health/low-vision/123>
- National Federation of the Blind
<https://nfb.org/>

- The Low Vision Gateway
<http://www.lowvision.org/>
- VisionAware: What is Low Vision?
<http://www.visionaware.org/info/your-eye-condition/eye-health/low-vision/123>

Scientific Articles on PubMed

- PubMed
<https://www.ncbi.nlm.nih.gov/pubmed?term=%28%28bradyopsia%5BTIAB%5D%29+OR+%28PERRS%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D>

OMIM

- PROLONGED ELECTRORETINAL RESPONSE SUPPRESSION
<http://omim.org/entry/608415>

Sources for This Summary

- Cheng JY, Luu CD, Yong VH, Mathur R, Aung T, Vithana EN. Bradyopsia in an Asian man. Arch Ophthalmol. 2007 Aug;125(8):1138-40.
Citation on PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/17698770>
- Hartong DT, Pott JW, Kooijman AC. Six patients with bradyopsia (slow vision): clinical features and course of the disease. Ophthalmology. 2007 Dec;114(12):2323-31. Epub 2007 Sep 12.
Citation on PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/17826834>
- Michaelides M, Li Z, Rana NA, Richardson EC, Hykin PG, Moore AT, Holder GE, Webster AR. Novel mutations and electrophysiologic findings in RGS9- and R9AP-associated retinal dysfunction (Bradyopsia). Ophthalmology. 2010 Jan;117(1):120-127.e1. doi: 10.1016/j.ophtha.2009.06.011. Epub 2009 Oct 8.
Citation on PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/19818506>
- Nishiguchi KM, Sandberg MA, Kooijman AC, Martemyanov KA, Pott JW, Hagstrom SA, Arshavsky VY, Berson EL, Dryja TP. Defects in RGS9 or its anchor protein R9AP in patients with slow photoreceptor deactivation. Nature. 2004 Jan 1;427(6969):75-8.
Citation on PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/14702087>
- Vincent A, Robson AG, Holder GE. Pathognomonic (diagnostic) ERGs. A review and update. Retina. 2013 Jan;33(1):5-12. doi: 10.1097/IAE.0b013e31827e2306. Review.
Citation on PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/23263253>

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